

ABSTRACT

Provided are methods and compositions for quantitatively measuring the amount of an unlabeled organic acid in a sample. Oxygen-18 labeled organic acids are used as internal standards to adjust for the loss of a structurally similar or identical unlabeled organic acid through processing required for its detection, such as by mass spectrometry. The methods of the invention are useful for diagnosing inborn errors of metabolism in an individual by quantitating signature organic acids in body fluids such as urine or plasma.